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Isolated absence of the right pulmonary artery with coexisting left-sided heart failure: case report and literature review

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ABSTRACT

Isolated unilateral absence of the pulmonary artery (UAPA) exhibits a benign course and often goes undiagnosed until adulthood. We present a 39-year-old male admitted for left-sided congestive heart failure (CHF) exacerbation. However, chest radiograph demonstrated findings suspicious for UAPA that was ultimately confirmed on computed tomography. Because both CHF and UAPA present similarly with exercise intolerance and dyspnea, a high index of suspicion is required to secure the diagnosis, and in this case, symptoms attributed to CHF may have contributed to delay. A strong awareness of typical radiographic findings allows for initiation of confirmatory tests necessary for a correct diagnosis.

1. Introduction

Unilateral absence of the pulmonary artery (UAPA) is a rare congenital anomaly that is frequently associated with other cardiac anomalies such as tetralogy of Fallot and a right aortic arch [1]. Most of these cases are diagnosed and surgically treated during infancy [2]. However, UAPA may also present as an isolated abnormality and often exhibits a relatively benign clinical course with minimal to absent or nonspecific symptoms [3]. As a result, isolated UAPA is commonly undiagnosed until adulthood when an incidental chest radiograph ordered for an unrelated reason often initiates a series of diagnostic tests to confirm the diagnosis. In this report, we present a case of isolated UAPA without associated congenital cardiac anomalies but with coexisting heart failure with reduced ejection fraction.

2. Case presentation

A 39-year old male presented to the emergency department with one day of acute onset dyspnea, orthopnea, and dry cough. Pertinent past medical history included recurrent respiratory tract infections, atrial fibrillation, pulmonary hypertension with a right ventricular systolic pressure of 53 mm Hg, left-sided heart failure with reduced ejection fraction of 35–40% reported in September 2014, and coronary artery disease status post stenting of the left anterior descending artery in 2013. Pertinent medications included simvastatin, carvedilol, furosemide, hydralazine, isosorbide dinitrate, aspirin, and warfarin. In the emergency department, he was found to be tachycardic with a heart rate of 150 with a 12-lead EKG demonstrating atrial flutter without ischemic changes, a blood pressure of 126/112, and required 15 L flow nonrebreather to maintain an oxygen saturation of 94%. Physical exam was notable for coarse breath sounds, a grade III/VI systolic ejection murmur, and lower extremity pitting edema. Laboratory findings were notable for a white count of 12.0, hematocrit of 47.2, hemoglobin of 15.7, creatinine of 1.24, AST of 148, ALT of 127, troponin of 0.080, BNP of 707, and total bilirubin of 2.0. A chest radiograph demonstrated an enlarged cardiac silhouette, splaying of the carina suggestive of left atrial dilation, vascular redistribution suggestive of pulmonary venous hypertension, and a right mid to lower lung airspace opacity (Fig. 1), thought to represent aspiration or lobar pneumonia. The patient was admitted and treated for congestive heart failure (CHF) exacerbation, rapid ventricular rate, which was presumably leading to decreased diastolic filling times and contributing to his dyspnea, and pneumonia. Not long after being transferred to the inpatient medicine service following admission, the patient was pulseless and unresponsive. Cardiopulmonary resuscitation was performed for approximately ten minutes followed by return of spontaneous circulation. The patient was intubated during this time and was subsequently transferred to the intensive care unit for further management.

On retrospective review of the chest radiograph, the right hilar shadow was not visualized and the right lung appeared hypoinflated in relation to the left, with left-to-right cardiomediastinal shift suggestive of volume loss (Fig. 1). A transthoracic echocardiogram was performed,

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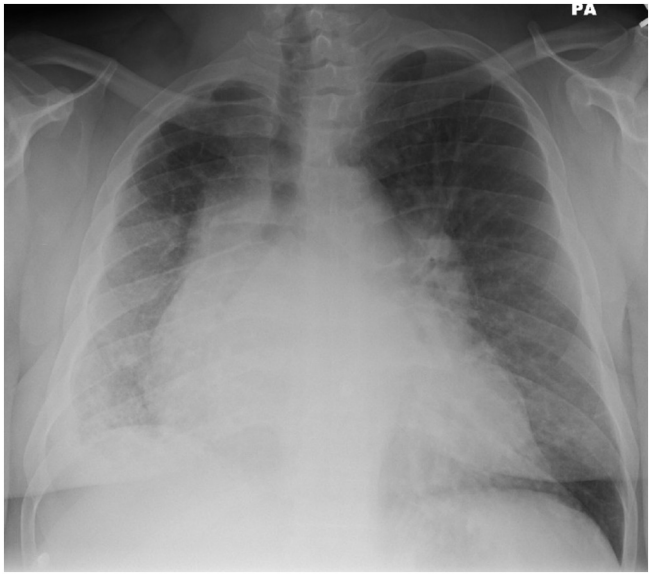

Fig. 1. A posterior-anterior frontal chest radiograph demonstrates an enlarged cardiac silhouette, splaying of the carina, and superolateral convexity of the cardiac contour along the left atrial appendage in keeping with left atrial dilatation. In addition, absence of the right hilar shadow, hypoinflated right lung, rightward cardiomediastinal shift, and a contralateral hyperinflated lung raise suspicion of an absent right pulmonary artery.

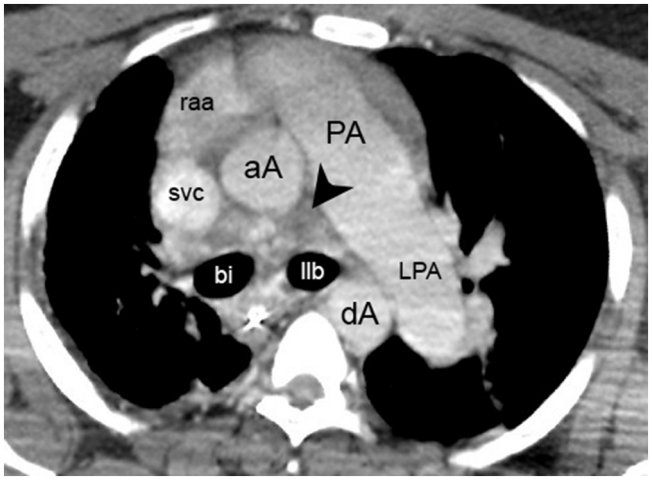

Fig. 2. Transverse contrast-enhanced chest CT image in the mediastinal window demonstrates the mildly dilated main pulmonary artery originating from the pulmonary outflow tract and traversing posterolaterally to become the left pulmonary artery. There is no visualization of a branch originating at this level to become the expected right pulmonary artery (black arrowhead), which is expected to traverse anterior to the right maintstem bronchus and bronchus intermedius. In addition, the is hypoinflation of the right lung and rightward cardiomediastinal shift as better characterized on CT. aA = ascending aorta; dA = descending aorta; PA = main pulmonary artery; LPA = left pulmonary artery; raa = right atrial appendage; svc = superior vena cava; bi = bronchus intermedius; llb = left lower lobe bronchus.

which demonstrated a moderately dilated left ventricle, an ejection fraction between 30 and 35%, a markedly dilated left atrium, mild pulmonary hypertension (41 mm Hg), and a dilated inferior vena cava with poor inspiratory collapse in keeping with elevated right atrial pressures. A computed tomography (CT) of the chest with intravenous contrast demonstrated a mildly dilated main pulmonary artery continuing as a dilated left pulmonary artery, and more distally as the left interlobar artery (Figs. 2–4). The right pulmonary artery was not visualized. CT of the chest also confirmed the presence of

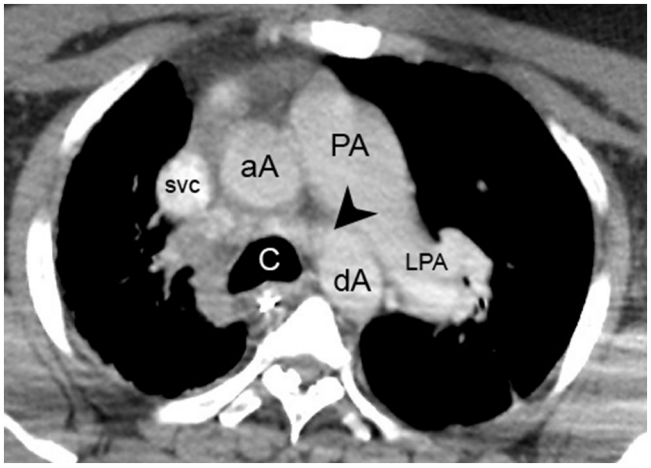

Fig. 3. On transverse contrast-enhanced chest CT image in the mediastinal window slightly more superior at the level of the carina, a dilated systemic collateral vessel originating from the anteromedial aspect of the descending thoracic aorta is suspected, and is seen traversing rightward and anterior to the carina to extend into the right hilum (black arrowhead). aA = ascending aorta; dA = descending aorta; PA = main pulmonary artery; LPA = left pulmonary artery; raa = right atrial appendage; svc = superior vena cava; c = carina.

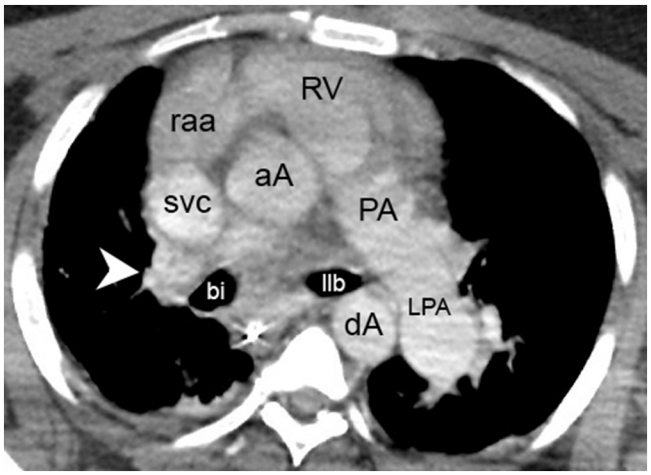

Fig. 4. More inferior at the level of the bronchus intermedius, the collateral vessel traverses anteriorly and laterally in relation to the bronchus intermedius, positioned in the expected location of the right interlobar artery (white arrowhead). dA = descending aorta; PA = main pulmonary artery; LPA = left pulmonary artery; raa = right atrial appendage; svc = superior vena cava; RV = right ventricle; bi = bronchus intermedius; llb = left lower lobe bronchus.

cardiomediastinal shift and better characterized the right lung volume loss previously seen on radiograph.

Following intubation and transfer to the intensive care unit, the patient became increasingly febrile as well as hypoxemic despite ventilator support. Despite treatment with broad-spectrum intravenous antibiotics, volume resuscitation and pressor support, he went on to develop severe sepsis, multi-organ failure, and septic shock. The patient developed pulseless electrical activity arrest, required cardiopulmonary resuscitation, and did not achieve return to spontaneous circulation. An autopsy was performed and was notable for edematous, hemorrhagic lungs with right lung hypoplasia. The cause of death was determined to be secondary to septic shock from pneumonia in the setting of underlying cardiac disease.

3. Discussion

Unilateral absence of the pulmonary artery (UAPA) is a rare congenital abnormality often associated with cardiac anomalies such as a right aortic arch, tetralogy of Fallot, patent ductus arteriosus, and septal defects [1], but may also be observed as an isolated abnormality [4]. Bouros et al. reported a prevalence of approximately 1 in 100,000 based on a review of 600,000 chest radiographs [2]. The anomaly is thought be due to the failure in connection between the sixth aortic arch and the pulmonary trunk during development [5]. While most congenital cases are diagnosed and treated during childhood [6], an isolated presentation of UAPA may go undetected, misdiagnosed, and exhibit a benign clinical course with minimal symptoms into adulthood [2], as seen in the current case. The literature does not appear to definitively establish a predilection for the left or right side [7], although Ten Harkel et al. and Kruzliak et al. in their literature reviews reported a higher incidence on the right [1,3]. Perfusion in the absence of a pulmonary artery often occurs through collaterals arising from the aorta and its branches, including the bronchial, subclavian, intercostal, and diaphragmatic arteries [8].

It has been reported that up to 30% of patients with isolated UAPA are asymptomatic [1,2,9]. Ten Harkel et al. reported hemoptysis in 20%, recurrent pulmonary infections in 37%, and limited exercise tolerance or dyspnea in 40% of patients. The cause of hemoptysis is thought to be related to excessive collateral circulation [3]. The pathogenesis of recurrent infections is less clear and is thought to be multifactorial. Alveolar hypocapnia leading to bronchoconstriction and mucus trapping, poor perfusion resulting in diminished delivery of inflammatory cells, and impaired ciliary clearance are hypothesized to be contributing factors to predispose to infections [3]. As seen in our case, pulmonary hypertension is common among patients with unilateral UAPA, and has ranged from 19% to 44% [1,10,11]. The pathogenesis is thought to be secondary to increased blood flow to the unilaterally present pulmonary artery, resulting in shear stress on the underlying endothelium, release of vasoconstrictive factors, and ultimately remodeling of the pulmonary vasculature that leads to increased resistance [3]. Interestingly, in contrast to expected changes related corpulmonale in the setting of pulmonary hypertension, our patient demonstrated a reduced left ventricular ejection fraction, left ventricular, and left atrial dilatation. It is difficult to determine if these symptoms were related to baseline CHF or to what extent the result of UAPA. Although the exact age of onset of CHF was not elucidated from the patient, the age of 39 is relatively young for heart failure, with Lloyd-Jones et al. reporting an incidence of 0.3% between the ages of 20–39, compared to 9.1% and 14.7% for those between the ages of 60–79 and 80 and above, respectively [12]. To our knowledge, our patient had no history of dilated cardiomyopathy and his CHF was attributed to his underlying coronary artery disease.

The diagnosis of UAPA can be easily missed or misdiagnosed without a high index of suspicion. Physical exam may reveal a systolic ejection murmur across the pulmonary outflow tract [3], as observed in the current case. Chest radiographs may demonstrate a smaller than normal hyperlucent lung, cardiomediastinal shift to the affected side, ipsilateral hemidiaphragmatic elevation, absent hilar shadow, and a contralateral hyperinflated lung [3]. A sufficiently high index of suspicion on radiographs will allow for further workup with cross-sectional imaging in which the diagnosis of UAPA can be definitively secured with contrast-enhanced CT of the chest. On CT, the absent pulmonary artery typically terminates within 1 cm from the origin of the main pulmonary artery. Ancillary findings may include a mosaic perfusion pattern likely due to small vessel disease and underlying pulmonary hypertension, dilated collateral vessels from the systemic circulation, and bronchiectasis from recurrent infections [3]. Transthoracic echocardiography may be utilized in excluding additional cardiac or vascular abnormalities and evaluating for pulmonary hypertension. Pulmonary angiography remains the gold-standard but is rarely performed unless embolization is required [3].

There remains to be a strong consensus on the treatment of isolated UAPA. Pharmacologic management aimed at treating pulmonary hypertension with prostacyclin, endothelin receptor antagonists, and calcium channel blockers have been reported with variable success [3]. Surgical options include pneumonectomy and revascularization of intrapulmonary arteries to the central pulmonary artery. Embolization may be used to treat massive hemoptysis. In our case, the patient was deemed a poor candidate for surgical and interventional treatment given his underlying septic shock, hemodynamic instability, and multiorgan failure.

In summary, given the relatively benign and inconspicuous clinical course of isolated UAPA, a high index of suspicion is required to make the diagnosis. A delay in diagnosis to adulthood in our case may have been partly attributed by the relatively indistinguishable cardiopulmonary signs and symptoms and exercise intolerance related to his left-sided CHF, which are similarly seen in isolated UAPA. Recognizing findings on chest radiograph that are often incidentally ordered for other indications is often key in initiating a series of appropriate diagnostic and confirmatory tests in ultimately securing the diagnosis.

Declaration of Competing Interest

The authors declare no conflicts of interests.

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